

Tolosa Hunt syndrome as initial presentation of systemic lupus erythematosus

By Fathima Nilofar

Case Report

2

Tolosa Hunt syndrome as initial presentation of systemic lupus erythematosus

Fathima Nilofar, Kavitha M M, Mahendra Kumar

Corresponding author:

Fathima Nilofar

E-mail: nilofarfathima14@gmail.com

ABSTRACT

A 54-year-old female patient with no known co-morbidities is the subject of this case study, who presented with Tolosa Hunt Syndrome (THS) as the first sign of Systemic Lupus Erythematosus (SLE). The patient complained of unilateral right-sided headache, double vision that appeared suddenly and right-sided eye discomfort. Surprisingly, there were no documented symptoms of vomiting or redness in the eyes, nor any complaints of vision loss, fever, stiff neck, or trauma. All other neurological examination were normal, but a clinical examination indicated right oculomotor nerve palsy, especially an inability to adduct, raise, or depress her right eye. Detailed clinical examination revealed alopecia areata, erythematous macular lesions on her earlobe. Lab investigations unremarkable except an increased erythrocyte sedimentation rate (ESR). Asymmetric thickening and augmentation of the right cavernous sinus on an MRI of the brain and orbit validated the diagnosis of THS. Subsequent laboratory testing revealed a speckled pattern with 4+ positive in ANA testing. Pulse therapy was initiated followed by oral steroids, hydroxychloroquine and mycophenolate mofetil. Patient improved and there were no relapses during routine follow-ups. This instance highlights the significance of considering autoimmune diseases, such as SLE, when dealing with unusual presentations of THS. Clinical signs, neuroimaging tests, and the patient's reaction to steroids were used to make the diagnosis. Laboratory and CSF tests confirmed the diagnosis by ruling out other ophthalmoplegia causes. Tissue biopsies are still the gold standard for diagnosis, but they should only be used as a last resort due to their high risk and intricate technical requirements. Finally, this research clarifies the uncommon co-occurrence of SLE and THS, highlighting the importance of thorough assessment and clinical attention in detecting difficult patients.

INTRODUCTION

The painful condition known as Tolosa Hunt Syndrome (THS) predominantly affects the cranial nerves and is typified by ophthalmoplegia. It is extremely uncommon for THS and SLE to coexist. This case report sheds light on the uncommon presentation of THS as a patient's first sign of SLE, highlighting the need for clinical vigilance in identifying complicated autoimmune diseases involving the cranial nerve.

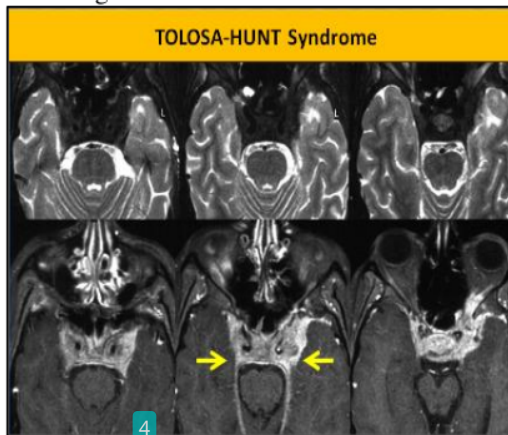


Figure 1: TOLOSA-HUNT syndrome

CASE PRESENTATION

The patient, a 54-year-old female, presented with complaints of double vision, headache persisting for a week, and eye pain over the past two days. Notably, she had no prior history of co-morbidities. The symptoms had varying durations, with the double vision persisting for two days, the headache for a week, and the eye pain for two days. Crucially, the absence of symptoms such as vision loss, fever, neck stiffness, trauma history, eye redness, and vomiting was noted. Clinical examination revealed right oculomotor nerve palsy, resulting in the inability to adduct, elevate, or depress the right eye, while the 4th and 6th cranial nerves appeared intact. Other neurological examinations yielded normal results.

INVESTIGATIONS

The investigations yielded notable findings across various parameters. Haematological examination indicated an elevated ESR of 84mm at 1 hour, suggesting an inflammatory response. Additionally, distinctive clinical manifestations included erythematous macular lesions on the earlobe and alopecia areata on the back of the scalp. Laboratory tests, particularly the ANA (Antinuclear Antibody) testing, revealed a speckled pattern with 4+ positivity, indicative of autoimmune activity. CSF (Cerebrospinal Fluid) analysis found normal. Neuroimaging through MRI of the Brain and Orbit provided critical insights, identifying asymmetric thickening and enhancement of the right cavernous sinus. These findings align with the clinical profile consistent with Tolosa Hunt Syndrome, a rare disorder characterized by inflammation of the cavernous sinus.

Important discoveries from the investigation helped to diagnose the patient's ailment. Alopecia areata on the scalp and erythematous macular lesions on the earlobe further indicated the possibility of an underlying autoimmune component [3]. Antinuclear Antibody (ANA) testing, in particular, showed a characteristic speckled pattern with a 4+ positive, suggesting an autoimmune process. MRI neuroimaging confirmed the diagnosis, showing asymmetric elevation and thickness of the right cavernous sinus a characteristic of Tolosa Hunt Syndrome (THS). Together, these studies yielded crucial information that helped with the precise diagnosis and patient care that followed.

DIAGNOSIS

After a thorough evaluation of the patient's clinical presentation and investigative results, Tolosa Hunt Syndrome (THS) was

determined to be the patient's diagnosis. In addition to a specific right oculomotor nerve palsy, the clinical symptoms were unilateral headache, eye discomfort, sudden-onset double vision, and horizontal diplopia [4]. Notably, THS is characterized by painful ophthalmoplegia and mostly affects cranial nerves. The fact that Systemic Lupus Erythematosus (SLE), a rare and complicated autoimmune disease, first manifested itself at THS makes this case unique. The extremely rare incidence of THS and SLE highlights the complexity of autoimmune diseases and the significance of identifying unusual clinical presentations [5]. This particular aspect of the case suggests that autoimmune diseases such as SLE should be taken into account when a patient presents with unusual symptoms of THS.

Understanding this relationship is crucial since it informs diagnostic and treatment decisions. It emphasizes the necessity of a careful and diligent examination when dealing with situations involving the involvement of the cranial nerve and implies that complicated autoimmune diseases might show up in strange ways [6].

The patient's treatment regimen comprised various interventions aimed at managing autoimmune symptoms effectively. Pulse Steroid Therapy was initiated, involving the administration of injectable methylprednisolone (1 gm IV) once a day for three days, with the primary goal of rapidly reducing inflammation and alleviating symptoms. Following this, oral steroids were introduced and tapered accordingly. Additionally, Hydroxychloroquine, a disease-modifying antirheumatic medication (DMARD), was administered to control inflammation associated with autoimmune conditions. The patient's treatment plan also incorporated Mycophenolate Mofetil, an immunosuppressive drug commonly used to regulate the immune system and curb further autoimmune activity; however,

specific details regarding the frequency of administration and dosage were not provided in the report [7][8].

The patient was receiving therapy for Tolosa Hunt Syndrome (THS) and its possible association with Systemic Lupus Erythematosus (SLE) using a comprehensive approach. This multimodal strategy addressed both THS and SLE, recognizing the case's complexity. The healthcare professional would customize the dosages and lengths of therapy based on the patient's reaction and general health.

The patient, following an extended period of medication and pulse steroid therapy, exhibited a significant improvement in symptoms associated with Tolosa Hunt Syndrome (THS). Remarkably, there were no relapses in THS symptoms during routine follow-up consultations, indicating the sustained efficacy of the prescribed treatment. The positive response to the recommended course of therapy underscores the effectiveness of the selected medications in managing both the primary symptoms of THS and potential underlying autoimmune causes. To ensure the ongoing stability of the patient's condition and to make any necessary adjustments to the treatment plan, continuous monitoring and follow-ups were diligently maintained.

This encouraging reaction emphasizes how crucial it is to treat Tolosa Hunt Syndrome (THS) patients appropriately and as soon as possible, as well as other autoimmune disorders.

DISCUSSION

The overall analysis of the case study emphasizes the remarkable combination of Tolosa Hunt Syndrome (THS) and Systemic Lupus Erythematosus (SLE), underscoring the complexity of autoimmune conditions and the significance of meticulous clinical

assessment. THS is an uncommon medical condition mostly affecting the cranial nerves that is characterized by uncomfortable ophthalmoplegia. In this case, the inflammatory illness SLE, which has a broad spectrum of clinical signs, presented unintentionally as its initial sign. This type of co-occurrence is quite rare and serves as an upsetting reminder of the broad spectrum of expressions autoimmune conditions may have. The multiple modalities used to treat the individual comprised hydroxychloroquine, mycophenolate mofetil and long-term oral steroids.

In addition to highlighting the need for an extensive medical evaluation when brain involvement is apparent, the present study emphasizes the need to consider autoimmune disorders like SLE when THS symptoms are atypical. It is a vital addition to the knowledge of complicated autoimmune illnesses, guiding both diagnosis and therapy in these challenging cases.

CONCLUSION

This case report sheds light on the uncommon confluence of Systemic Lupus Erythematosus (SLE) and Tolosa Hunt Syndrome (THS). The example highlights how autoimmune diseases can appear in a variety of ways clinically and the importance of a thorough examination in complicated situations including cranial nerve damage. The effectiveness of the selected therapeutic technique is demonstrated by the patient's favorable reaction to therapy, which is characterized by clinical improvement and the lack of relapses. This unusual instance emphasizes the need for caution when identifying unusual THS symptoms and implies that autoimmune diseases should be taken into account when handling unusual presentations. advances knowledge of autoimmune disorders.

FIGURE 1

ERYTHEMATOUS MACULAR
LESIONS IN EAR LOBE



FIGURE 2
SPECKLED PATTERN WITH 4+
POSITIVITY

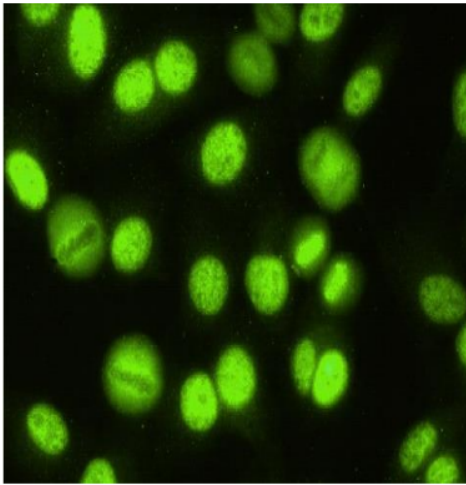
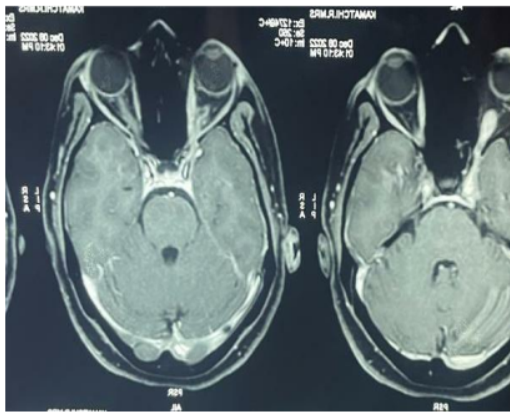


FIGURE 3

MRI Brain and Orbit done showed asymmetric thickening and enhancement of right cavernous sinus consistent with Tolosa Hunt Syndrome



REFERENCES

- [1] Srinivasan A, Milman T, Lane KA, Bilyk JR. Pathology of the Orbit: Inflammations and Infections. Albert and Jakobiec's Principles and Practice of Ophthalmology. 2020:1-47.
- [2] Lapić I, Padoan A, Bozzato D, Plebani M. Erythrocyte sedimentation rate and C-reactive protein in acute inflammation: meta-analysis of diagnostic accuracy studies. American journal of clinical pathology. 2020 Jan 1;153(1):14-29.
- [3] Trüeb RM, Gavazzoni Dias MF, Dutra Rezende H, de la Cruz Vargas Vilte RM, Romiti R. The Hair and Scalp in Systemic Infectious Disease. In Hair in Infectious Disease: Recognition, Treatment, and Prevention 2023 Aug 13 (pp. 303-365). Cham: Springer International Publishing.
- [4] Syed SB, Mourra AA, Chatterjee T. Isolated Unilateral Abducens Nerve Palsy Manifesting as a Rare Complication of Idiopathic Pituitary Apoplexy: A Case Report. Cureus. 2022 Feb 20;14(2).
- [5] Harley IT, Sawalha AH. Systemic lupus erythematosus is a genetic disease. Clinical Immunology. 2022 Mar 1;236:108953.
- [6] Dolezal O. Clinical Cases in Neurology. Springer; 2019 May 29.
- [7] Dima A, Jurcut C, Arnaud L. Hydroxychloroquine in systemic and autoimmune diseases: Where are we now? Joint Bone Spine. 2021 May 1;88(3):105143.
- [8] Bhat R, Tonutti A, Timilsina S, Selmi C, Gershwin ME. Perspectives on Mycophenolate Mofetil in the Management of Autoimmunity. Clinical Reviews in Allergy & Immunology. 2023 Jun 20:1-5.
- [9] Young SM, Lim AY, Lang SS, Lee KO, Sundar G. Efficacy and safety of pulsed intravenous methylprednisolone in early active thyroid eye disease. Orbit. 2019 Sep 3;38(5):362-9.
- [10] Schrezenmeier E, Dörner T. Mechanisms of action of

hydroxychloroquine and chloroquine:
implications for rheumatology. *Nature
Reviews Rheumatology*. 2020
Mar;16(3):155-66.